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Congenital dacryocystocele: five clinical cases

C. Hupin, N. Lévèque*, Ph. Eloy**, B. Bertrand** and Ph. Rombaux**

*Department of Otorhinolaryngology and Head and Neck Surgery, University of Louvain, Cliniques Saint Luc, Brussels, Belgium; **Department of Otorhinolaryngology and Head and Neck Surgery, University of Louvain, Cliniques de Mont-Godinne, Yvoir, Belgium

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Abstract. *Congenital dacryocystocele: five clinical cases.* Congenital dacryocystocele (CDC) is recognised as a cause of nasal airway obstruction or respiratory distress in newborns. CDC is caused by the distal obstruction of the lachrymal duct and presents as a cystic formation in the inferior meatus.
We discuss five cases of dacryocystocele, together with surgical management and outcome. Endoscopic endonasal marsupialisation and appropriate postoperative care resulted in definitive recovery for all patients.
In newborns or infants with nasal obstruction, CDC should be considered in the differential diagnosis, and prompt endoscopic endonasal marsupialisation is mandatory.

Introduction

Congenital dacryocystocele (CDC) is one of the causes of nasal obstruction in newborns. It presents as a nasolachrymal cyst or obstruction of the nasolachrymal duct (NLD) with cystic extension into the nasal fossa. When tears and secretions accumulate in the lachrymal sac as a result of an upper or lower obstruction in the lachrymal drainage apparatus, there is dilatation of the lachrymal sac and often protrusion of the cyst into the nasal lumen.[1]

Dacryocystocele, also called dacryocele, lachrymal sac cyst, nasolachrymal duct mucocele, amniotocele or amniocele is a rare disease.[2] Nevertheless, this condition must be included in the differential diagnosis of any newborn or infant with respiratory distress and/or nasal obstruction.

We describe five cases of neonates with CDC. Patients presented either with nasal obstruction, epiphora or a bluish swelling in the region of the medial canthus and a cystic formation in the inferior meatus.

In each case, we performed endoscopic endonasal marsupialisation, followed by appropriate postoperative care, resulting in recovery for all patients. No recurrence was observed during follow-up.

Case Report

Case 1

A full-term male infant presented with dyspnoea, with evidence of nasal obstruction, immediately after birth. Choanal atresia and pyriform aperture stenosis were excluded after clinical examination. Purulent rhinitis was diagnosed, with Streptococcus Viridans. Local care was prescribed: saline solution rinses, oxymetazoline, bacitracine-neomycine. The infant was discharged after four days of life and evolution was excellent. Two weeks later, he was admitted to A&E for respiratory distress. The parents had stopped the local care 24 hours previously. Physical examination showed he was tachypnoeic and using accessory muscles of respiration. Bilateral sibilant rales and purulent nasal discharge were noted, without fever. Nasal fibroscopy showed a cystic mass filling the right inferior meatus.

The patient was taken to the operating room with the diagnosis of right dacryocystocele. Nasal endoscopy under general anaesthesia confirmed the diagnosis and marsupialisation was performed, in conjunction with probing of the lachrymonasal duct. A peroperative CT scan showed a distended lachrymal sac and NLD.

Nasal breathing improved immediately after surgery and the patient remained asymptomatic during the three years of follow-up.

Case 2

An 8-day-old male infant was referred to our department for the

evaluation and treatment of a right periorbital abscess.

Urinoma due to a junction syndrome was diagnosed during pregnancy, with good spontaneous evolution. Biometrics were normal. Birth at term via spontaneous vaginal delivery was uncomplicated. The Apgar score was normal. Over the course of the next day, a bilateral purulent ocular discharge was seen. Topical antibiotics (ofloxacine) were administered. Two days later, periorbital erythema and upper lid swelling appeared on the right side. The patient was given oral antibiotics (amoxicillin-clavulanic acid + erythromycin) and the symptoms improved for 48 hours, after which the swelling increased again, as did the ocular discharge. IV antibiotics were started (amoxicillin-clavulanic acid + netilmicine), and oral erythromycin and local ofloxacine were maintained. The day after, exophthalmia was noted and so a CT scan was performed. It showed a right periorbital abscess. No fever was observed throughout this period.

Six days after the onset of the first symptoms, the infant was transferred to our hospital. Clinical examination showed right exophthalmia, major palpebral swelling, purulent ocular discharge and decreased ocular horizontal motility. The diagnosis of right NLD obstruction with dacryocystitis, periorbital cellulitis and subperiosteal abscess was made. IV antibiotics were changed to IV cefotaxime + oral erythromycin. Surgery was scheduled for the same day. Endoscopy-guided marsupialisation was performed, as well as ethmoidectomy and drainage of the subperiosteal abscess. Three millilitres of purulent material were expressed. No organisms grew in any cultures of this discharge. Evolution was rapidly positive, with reduction of the periorbital swelling and normalisation of ocular motility. Postoperative lachrymal sac massages as well as saline rinses were performed. At present, no recurrence has been observed.

Case 3

A 2-week-old male infant whose parents and neonatologists had noted slow progressive respiratory distress associated with nasal obstruction was transferred to our hospital. The medical history revealed an uncomplicated pregnancy, gastro-oesophageal reflux since birth and neonatal jaundice treated with five days of phototherapy. Clinical examination upon admission showed right conjunctivitis related to an infected homolateral lachrymal cyst. Purulent discharge was present, which grew Streptococcus Viridans and Staphylococcus (coagulase-negative) on culture. Nasal endoscopy was performed and bilateral lachrymal cysts with right dacryocystitis were found (Figures 1a,1b). The patient was given intravenous antibiotics with anti-reflux treatment and local care (topical antibiotics and vasoconstrictor nasal drops). Forty-eight hours later, the lachrymal cysts were marsupialised under endoscopic control. Following surgery, a marked improvement was seen in the patency of the nasal airways and respiratory difficulties improved immediately.

Two days later, the patient was discharged, with oral amoxicillin-clavulanic acid, oxymetazoline, saline rinses and massage of the internal canthus. During follow-up (16 months at present), there has been no evidence of recurrence.

Case 4

A two-month-old male infant was referred to our ENT outpatient clinic for the evaluation of left epiphora. Since birth, the patient had presented a left ocular discharge. Pressure on the medial canthus showed pus in the lachrymal points. Probing had been performed previously under local anaesthesia without success. Clinical examination showed a bluish-grey cystic swelling arising beneath the left inferior turbinate. Diagnosis of left CDC was confirmed.

Endonasal endoscopic marsupialisation resulted in complete resolution of the mucocele and improvement in symptoms. Microbiology revealed Staphylococcus (coagulase-negative). The patient remained asymptomatic during the 6 months of follow-up.

Case 5

A 12-day-old female infant with uneventful birth and pregnancy was referred to our ENT outpatient clinic with a diagnosis of bilateral dacryocystocele. The infant was born via spontaneous vaginal delivery. At birth, facial swelling was noted by parents and nurses. The swelling worsened progressively, especially at the internal angle of the orbit. Respiratory distress was noted, especially during feeding. Choanal atresia had been excluded by passing an aspiration sonde through both nostrils. The infant had been examined by an ENT specialist who had diagnosed bilateral CDC and referred her to

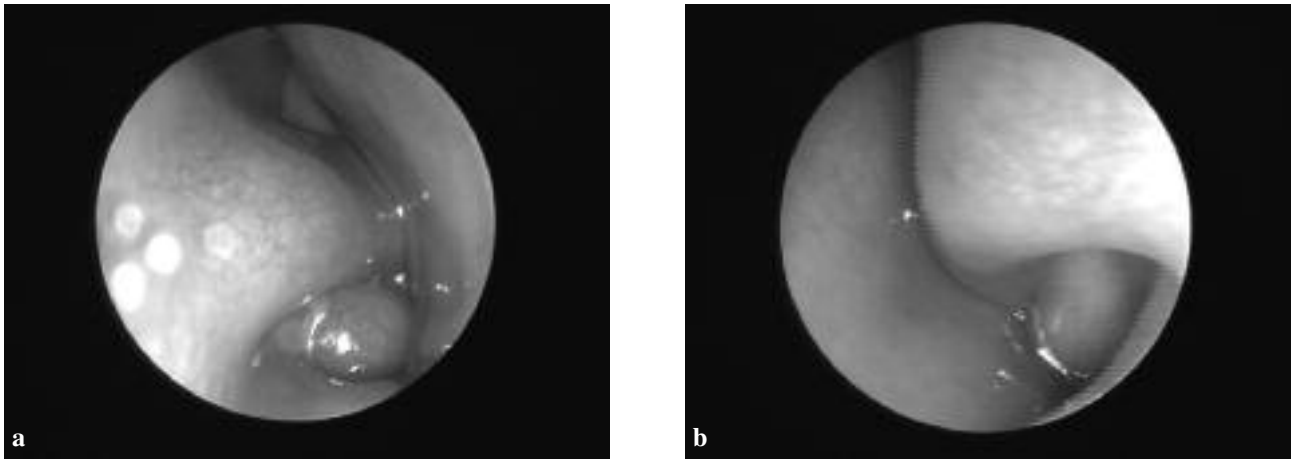

Figure 1
Endoscopic endonasal view of the inferior meatus, showing a cyst beneath the inferior turbinate. a: right nasal fossa; b: left nasal fossa

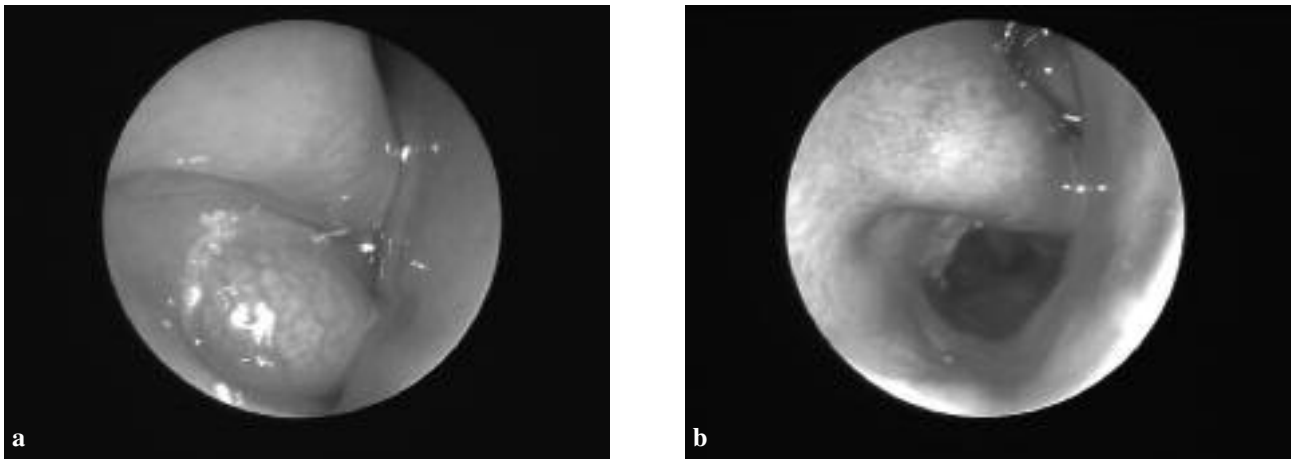

Figure 2
Endoscopic endonasal view of the right inferior meatus with cyst formation. a: before marsupialisation; b: after marsupialisation

our hospital. Clinical examination revealed a bluish cutaneous mass inferior and lateral to the lachrymal sac, with hypertelorism, caused by the extra soft tissue medial to the eye. Nasal endoscopy showed cystic masses filling the inferior meatus of both nasal cavities, confirming the diagnosis.

At 20 days of life, under direct endoscopic visualisation, the cyst walls were marsupialised and a thick discharge was evacuated (Figures 2a,2b). Just before starting surgery, we took advantage of the general anaesthesia to perform a CT scan of the head and sinus (to exclude other causes for the hypertelorism), which showed bilateral cystic dilatation of the lachrymal sac and nasolachrymal duct, forming masses that obstructed both inferior meatus (Figures 3a,3b,3c,3d). No other pathologic findings were detected. A histological section of the cyst showed normal nasal mucosa. Immediately after surgery, periorbital and facial swelling decreased and respiratory symptoms improved significantly. The day after, patient was sent home, with oral amoxicillin-clavulanic acid, local oxymetazoline, saline rinses and internal canthus massages. At present, no recurrence has been noted.

The summary of the clinical characteristics of these five patients is presented in Table 1.

Discussion

CDC is an uncommon disease which is mainly caused by an obstruction at the distal end of the nasolachrymal duct.[3]

Table 1
Clinical characteristics of CDC

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Gender	Male	Male	Male	Male	Female
Age at diagnosis	18 days	8 days	14 days	2 months	2 days
Laterality	Right	Right	Bilateral	Left	Bilateral
Nasal obstruction	+	+	+	+	+
Respiratory distress	+	-	+/-	-	-
Complications	None	Periorbital cellulitis	Dacryocystitis	None	None
Other congenital anomalies	None	None	None	None	None
Follow-up	38 months	45 months	16 months	6 months	2 months

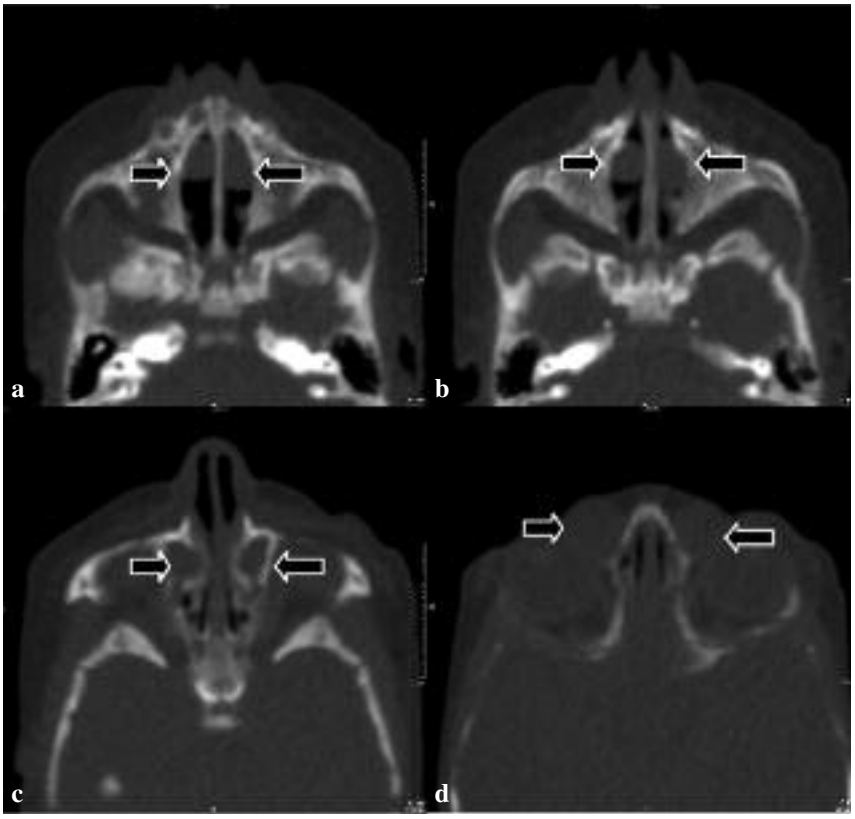

Figure 3
CT scan in horizontal planes, showing a cystic formation in the inferior meatus (right and left) and increased volume of lachrymal sac due to obstruction of the lachrymal pathways. Arrows indicate dacryocystocele in the inferior meatus. 3a,b,c,d: from below to upper part of the lachrymal pathways.

Symptoms of CDC can be present at birth or appear a few days later, as in two of our cases. This pathology is more frequent in females (65 to 80%) and can be either uni- or bilateral.[1,2,4] Only one of our patients was female and three presented with unilateral disease.

The disease usually presents with an intranasal cystic mass in the inferior meatus, frequently associated with epiphora and/or a painless bluish swelling below the medial canthus.[5] Complications such as dacryocystitis, preseptal cellulitis, and respiratory distress have been described.[4,6]

In newborns with respiratory distress and/or nasal obstruction, differential diagnosis should consider several diseases: malformations include choanal atresia, pyriform aperture stenosis, craniofacial dysmorphia and CDC.[7,8] It should be noted that upper airway obstruction secondary to CDC did not usually lead to severe respiratory distress requiring intubation or intensive care. Tumours should be excluded (teratoma, nasal glioma, encephalocele, meningocele, hemangioma) as well as infectious (neonatal rhinitis) or traumatic aetiologies (septal injury during birth).[9]

The diagnosis of CDC is made by clinical observation. A complete clinical examination is therefore essential for a correct diagnosis and to provide appropriate

treatment. Facial examination, anterior rhinoscopy with palpation of the cyst and meticulous nasal endoscopy are mandatory. If necessary, further investigations may include facial computed tomography scan or MRI. Imaging may help in the evaluation of other craniofacial abnormalities.

Management of CDC requires conservative and surgical management.[10]

Conservative treatment may be advised in uncomplicated cases[2,6] and includes nasal decongestion, saline rinses of nasal fossae and lachrymal ducts, topical antibiotics and lachrymal sac massage. This may lead to a spontaneous opening of the inferior meatus of the lachrymal duct but is not successful in the vast majority of the cases.

We recommend the early consideration of surgical management to avoid complications such as infection (dacryocystitis, cellulitis) or respiratory difficulties. Typically, under general anaesthesia, drainage of the cystic mass is performed, followed by marsupialisation of the cyst itself through an endoscopic endonasal approach. The introduction of a bicanaliculonasal probe in the lachrymal duct is not mandatory in primary cases.

Surgical procedure starts with vasoconstrictive agents (cottonoid pledgets soaked in a solution of epinephrine: one gamma/kg). The endoscopes used were 2.7 mm and 4 mm in diameter, 0° or 30° (Storz-Hopkins telescopes) clipped to a camera. After confirming diagnosis, the CDC was opened using a straight Blakesley forceps. Resection of the inferior mucosa helped to marsupialise the CDC and the contained secretions were suctioned. Care was taken to ensure a patent opening of the lachrymal pathway. No nasal packing was required except in the patient (2) where an ethmoidectomy was also performed.

Oral and topical antibiotics, normal saline solution in the nasal fossae and repeated massage of the lachrymal sac should be administrated after surgical procedure.

Conclusion

Congenital dacryocystocele is a rare clinical entity but all neonates with respiratory distress and/or nasal obstruction should be examined for this diagnosis. Anterior rhinoscopy and endonasal endoscopy with visualisation of the inferior nasal meatus are therefore required.

To facilitate nasal breathing and to avoid complications, we recommend prompt endoscopic endonasal marsupialisation of the cyst in CDC.

The current results indicate that surgical treatment is successful in all neonates/infants with early endonasal endoscopic surgery.

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Ph. Rombaux, M.D., Ph.D.
Department of Otorhinolaryngology and Head and Neck Surgery
University of Louvain
Cliniques Saint Luc
avenue Hippocrate 10
B-1200 Brussels, Belgium
Tel.: +32.2 764 19 30
Fax: +32.2 764 89 35
E-mail: philippe.rombaux@uclouvain.be